"A STUDY OF INTRACARDIAC REPAIR AND EFFECT OF TRANSANNULAR PATCHING ON RIGHT VENTRICULAR FUNCTION IN ADULT TETRALOGY OF FALLOT PATIENTS"

Dissertation submitted in partial fulfillment of
the requirement for the degree of

M. Ch. (Cardio Vascular & Thoracic Surgery)

BRANCH - I

MADRAS MEDICAL COLLEGE
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CHENNAI – 600 003

THE TAMIL NADU

DR. M.G.R MEDICAL UNIVERSITY

CHENNAI – 600 032

AUGUST 2010

CERTIFICATE

This is to certify that the dissertation entitled **"A STUDY OF INTRACARDIAC REPAIR AND EFFECT OF TRANSANNULAR PATCHING ON RIGHT VENTRICULAR FUNCTION IN ADULT TETRALOGY OF FALLO PATIENTS"** is the bonafide original work of **DR. R. MEENAKSHI SUNDARAM M.S.,** in partial fulfillment of the requirements for M.Ch. Branch-I CARDIO-VASCULAR & THORACIC SURGERY examination of THE TAMIL NADU DR. M.G.R MEDICAL UNIVERSITY to be held in August 2010. The period of post-graduate study and training was from August 2007 to July 2010.

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DECLARATION

I **Dr. R. MEENAKSHI SUNDARAM M.S.,** solemnly declare that this dissertation entitled, **"A STUDY OF INTRACARDIAC REPAIR AND EFFECT OF TRANSANNULAR PATCHING ON RIGHT VENTRICULAR FUNCTION IN ADULT TETRALOGY OF FALLO PATIENTS"** is a bonafide work done by me at the Department of Cardio Vascular & Thoracic Surgery, Madras Medical College and Government General Hospital during the period 2007 – 2010 under the guidance and supervision of the Professor and Head of the Department of Cardiothoracic Surgery, Madras Medical College and Government General Hospital, **Prof. S. Manoharan, M.S., M.Ch.** This dissertation is submitted to The Tamil Nadu Dr.M.G.R Medical University, towards partial fulfillment of requirement for the award of **M.Ch. Degree (Branch – I) in Cardio-vascular & Thoracic Surgery.**

Place : Chennai

Date: 24th MAY 2010

Dr. R. MEENAKSHI SUNDARAM

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CONTENTS

Tetralogy of fallot is the most common congenital cyanotic heart disease. Complete intracardiac repair of Tetralogy of Fallot (TOF) is being performed since 1950s. It constitutes about 10% of all congenital heart diseases. Current surgical management of TOF involves total correction in a single stage in the first six months of life. There are also adults who underwent palliative shunts in the childhood but never underwent the corrective procedure. In addition, in our part of the country a considerable number patients with well balanced TOF defect and adequate pulmonary stenosis to protect the pulmonary vasculature, will reach adulthood without any surgical intervention.

The first successful repair was performed by Lillehei and Varco in 1954 using controlled cross circulation. Kirklin reported the first repair using a pump oxygenator. Kirklin and others made important contributions to the surgical correction in the form of timing of operative repair, he indications for transannular patching to relieve Right Ventricular Outflow Tract(RVOT)obstruction and the use of grafts to reconstruct the RVOT. Today surgical repair beyond infancy is an exception rather the rule he poor natural history indicates that hemodynamic impairment in the survivors to adulthood may indicate the less severe spectrum of TOF Surgery after long standing cyanosis in

these subgroup of cases has caused controversy although several recent studies show better long term outcomes following surgery in this subset. This study present a three year followup of adult patients undergoing total correction for TOF in our institution.

1. To study the incidence and demography of Tetralogy of Fallot patients in the study population.

2. To study the symptomatology and mode of presentation

3. To study the coexisting anamolies

4. To study the efficacy of total correction in the TOF patients

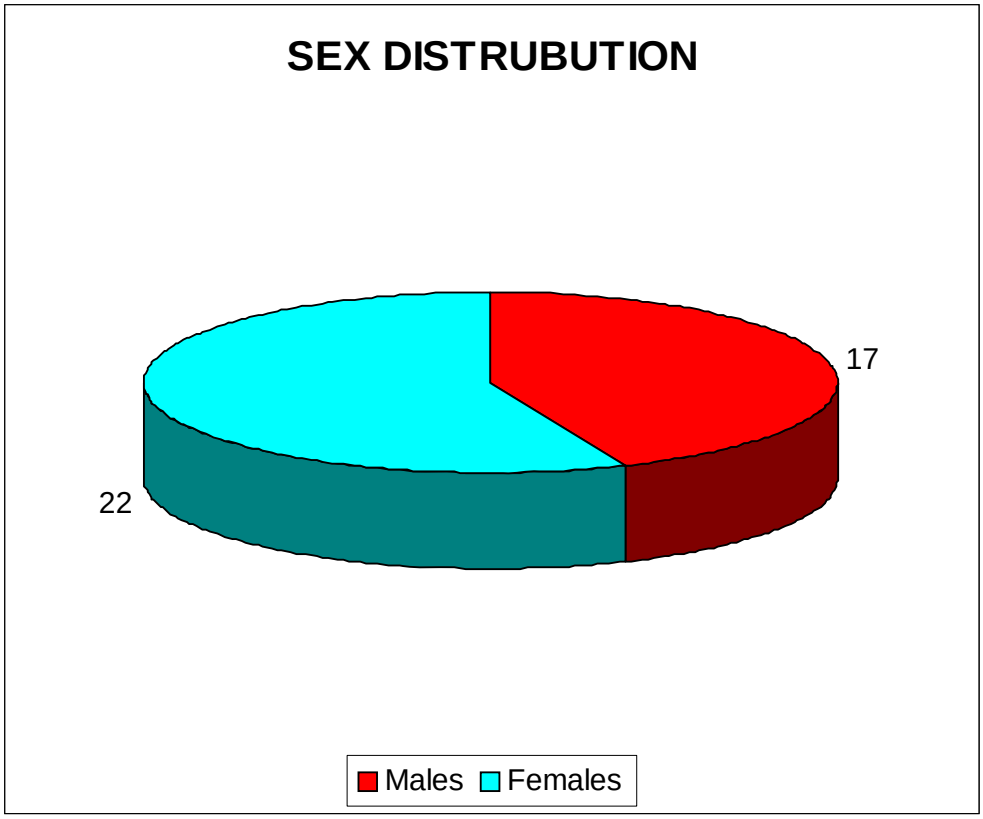
5. To study the effect of repair on the right ventricular function

6. To study the clinical status of patients and analye the hemodynamic data in the postoperative period.

7. To document a couple of interesting cases with unusual presentations

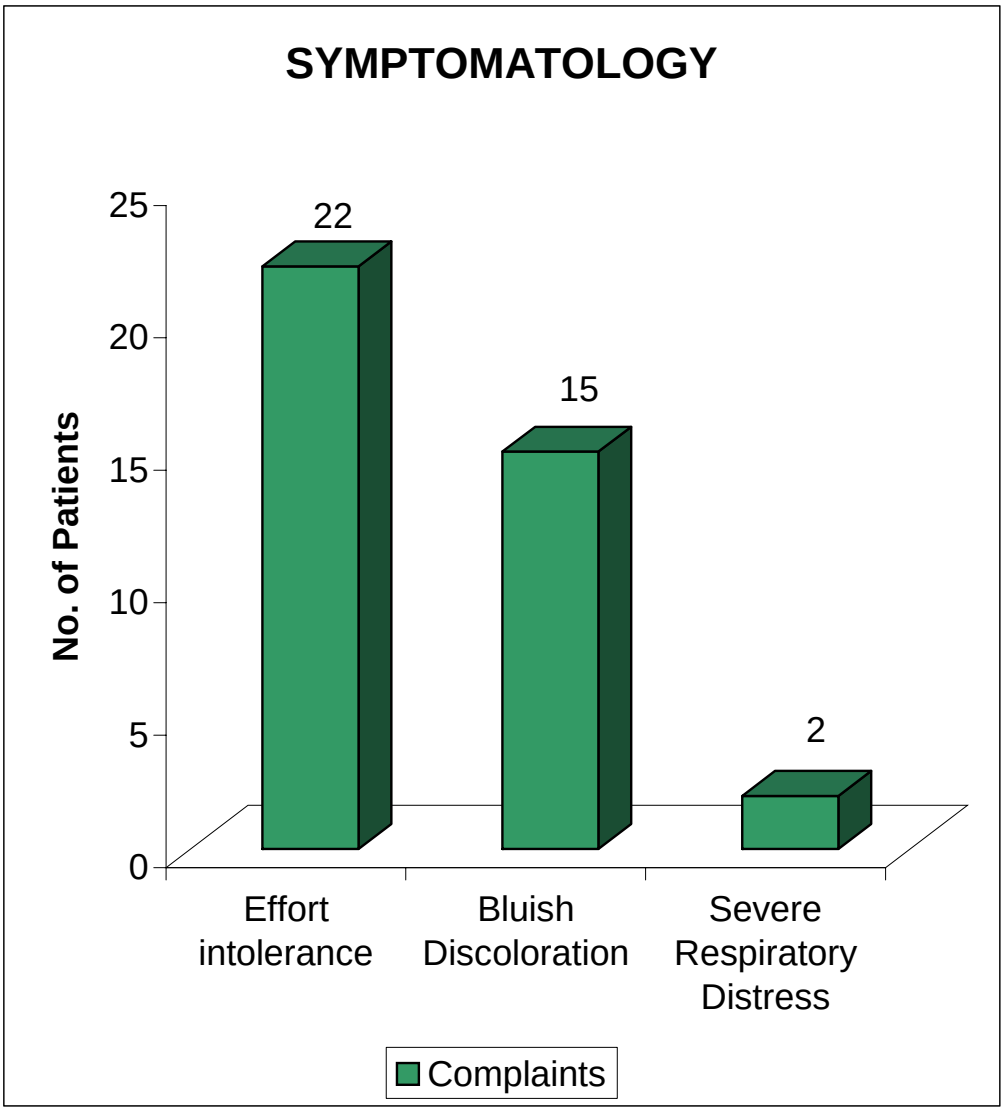
42 patients were admitted between August 2007 and April 2010 with a diagnosis of TOF. Of them 39 underwent intracardiac repair with or without transannular patching in the Department of Cardiothoracic Surgery, Government General Hospital, Chennai. We made a prospective study to assess the outcome of surgical correction in the study population during the study period and also reviewed the patients demographics, clinical status, morphology of RVOT, the different techniques used in the repair. We made an assessment of postoperative improvement of symptomatology and exercise tolerance. We made a periodic assessment of ventricular function and development of pulmonary regurgitation and residual pulmonary stenosis by echocardiography.

Between August 2007 and April 2010, 42 patients were admitted in the Deparment of Cardiothoracic Surgery, GGH, Chennai, with a diagnosis of TOF.39 patients underwent surgery.6 patients died in the immediate postoperative period.1patient died during the followup.Sex distribution 22 patients were males.17 patients were females.

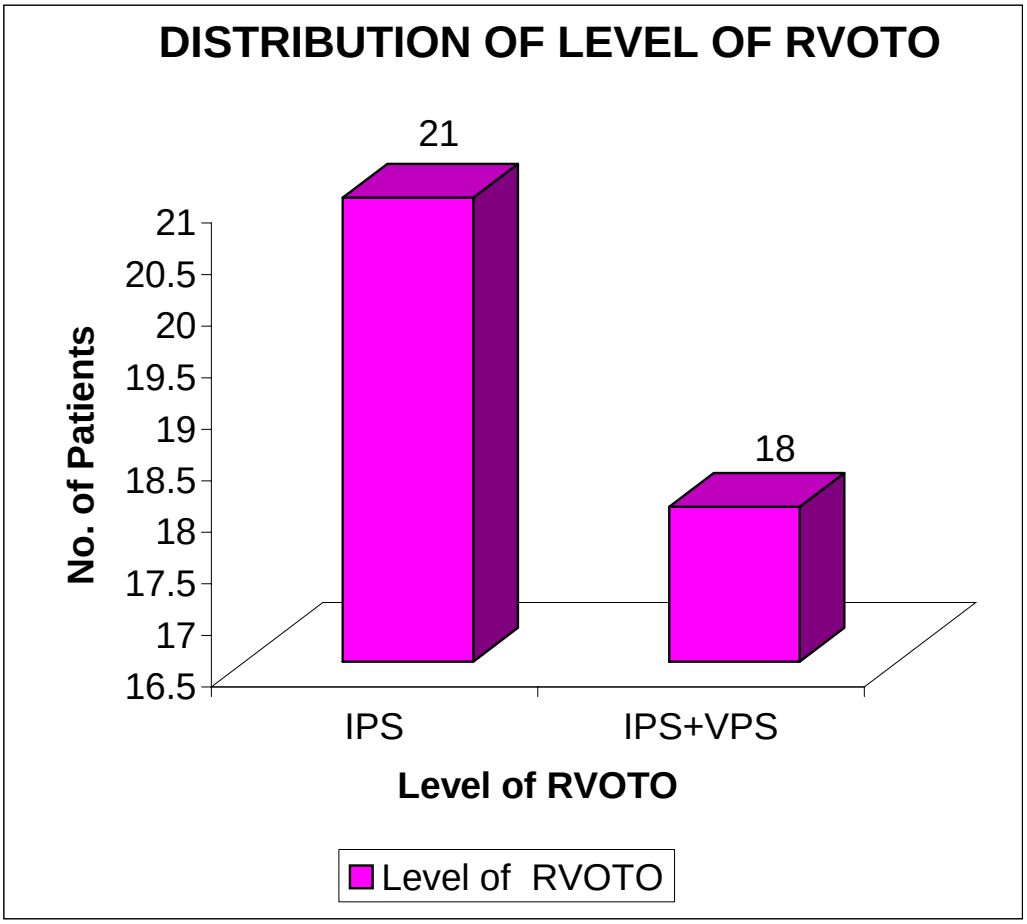


Age ranged from 11 to 27 years. The symptomatology of patients include exertional dyspnea, effort intolerance and bluish discoloration. Effort intolerance was the chief complaints in 65% of patients. Bluish discoloration was the chief complaint in 35% of cases.5patients were in

class 1 symptom.20 patients were in class 2 symptoms. 13 patients were in class 3 symptoms. One patient in class 4.



30% gave a positive history of squatting. One patient gave a history of brain abscess. One patient had a BT shunt. The preoperative hematocrit ranged between 40% to 68%.Cyanosis was not seen in 5 patients. One patient was a Downs syndrome. Preoperatively all patients were in sinus rhythm.Chest XRay revealed left aortic arch in all patients. Preoperative echocardiography revealed Infundibular Pulmonary Stenosis (IPS) in 55% cases. ValvarPulmonaryStenosis (VPS) and IPS in 45% cases.



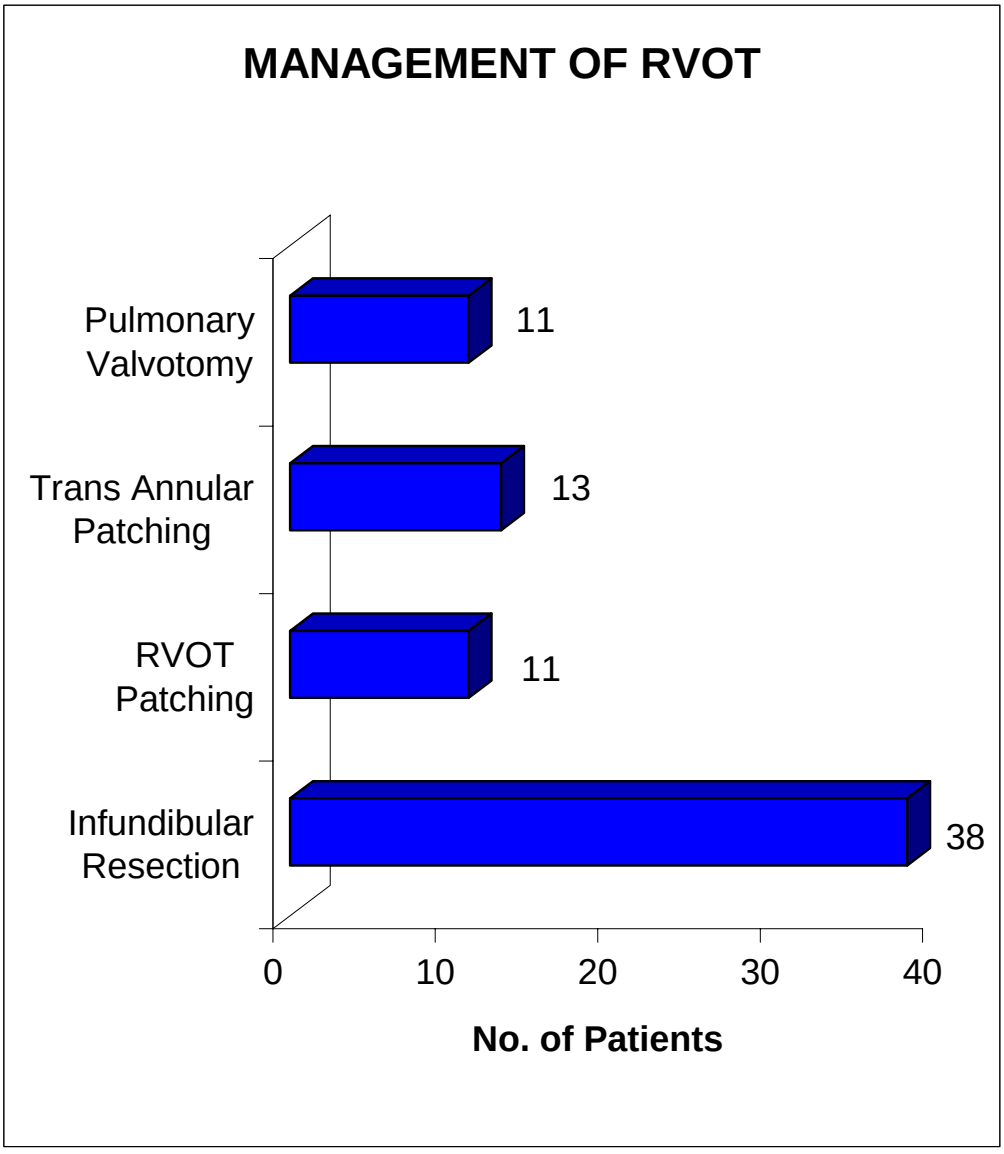
RVOT gradient was 55mmHg to 110mHg.Catherisation study was done in 30 patients. All patients had large subaortic VSD.All patients had preoperative evaluation of the size of pulmonary trunk, Right Pulmonary Artery(RPA), Left Pulmonary Artery (LPA), competency of pulmonary valve and coronary anatomy.Z value of the pulmonary annulus was identified preoperatively.

Surgical procedure

After median sternotomy on table assessment of size of MPA,RPA,LPA was made. All patients underwent total correction under Cardio Pulmonary Bypass (CPB),moderate hypothermia and cold blood cardioplegia. After crossclamping, through transatrial approach after retracting the septal leaflet of the tricuspid valve, VSD identified and was closed with Goretex patch with interrupted mattress sutures. Adequate resection of the muscle bands in the infundibulum was done through atrial incision. MPA was also opened to assess the adequacy of resection.

The required annulus size for the body surface was calculated based on the normograms and the corresponding size Hegar dilator was used to assess the adequacy of resection and the need for RVOT or

transannular patching. Where necessary pulmonary annulus was incised to achieve a relaxed dimension of 2-3mm larger than normal.RVOT/transannular patching was done with untreated autologous pericardium.



Pulmonary monocusp valve was not used in our study. RVOT patch was used in 11 patients and transannular patch was used in 13 patients.Intraoperative measurement of RV /LV pressures before and after repair was done. RVOT pressures at the infundibulum and MPA before and after repair were measured and compared to detect any residual gradients and RVOTO..RV/LV pressure ratio was between0.6 to 0.7.Length of crossclamp time ranged from 70-110 minutes and CPB time varied between 90-200 minutes. Postoperatively the duration of ICU stay, need for prolonged ventilatory support more than 48 hours and prolonged inotropic support were noted. Echocardiographic assessment of RV function and Pulmonary valve was done in the immediate postoperative period.

Follow up

The prospective evaluation of the patients was done by assessment of the clinical status and assessment of the RV function and competency of pulmonary valve, RV dimensions, tricuspid insufficiency by echocardiography. The echocardiographic evaluation of RV function was by Doppler imaging of the tricuspid annulus. The degree of pulmonary insufficiency or residual stenosis was assessed by a combination of qualitative and quantitative methods.Echocardiographic data were categorized as follows. RV dimension was categorized as

normal RV or dilated RV. Pulmonary regurgitation was categorized as mild,moderate,severe. Right ventricular function was recorded as RVEF. Right ventricular dysfunction and pulmonary valve insufficiency slowly develop after repair of tetralogy of Fallot and become present in almost all patients to some degree.Similar to Meijboom and coworkers , Zakha and associates and Horneffer and associates,who also used echocardiography in followup,we found evidence of more than trivial PR in a considerable number of patients. Exercise capacity is found to be decreased in patients with dilated RV, but most patients remain asymptomatic. Although some argue that long-standing pulmonary regurgitation has no adverse effect on survival , Pulmonary Valve Replacement will become a more frequent consideration as the population ages. In patients without operation and with anatomically normal hearts, symptoms of right heart failure will develop 30 to 40 years later in life. There were no reoperations during the study period.Nine patients required prolonged ventilation for more han 48 hours and prolonged inotropic support. Two patients underwent reexploration for bleeding. Two patients reexplored under suspicion of cardiac tamponade. On followup 20 patients were in class1, 13 patients were in class2.6 patients had mild to moderate PR,8 had moderate PR, 3 severe PR in echocardiography. (Tabl 1).

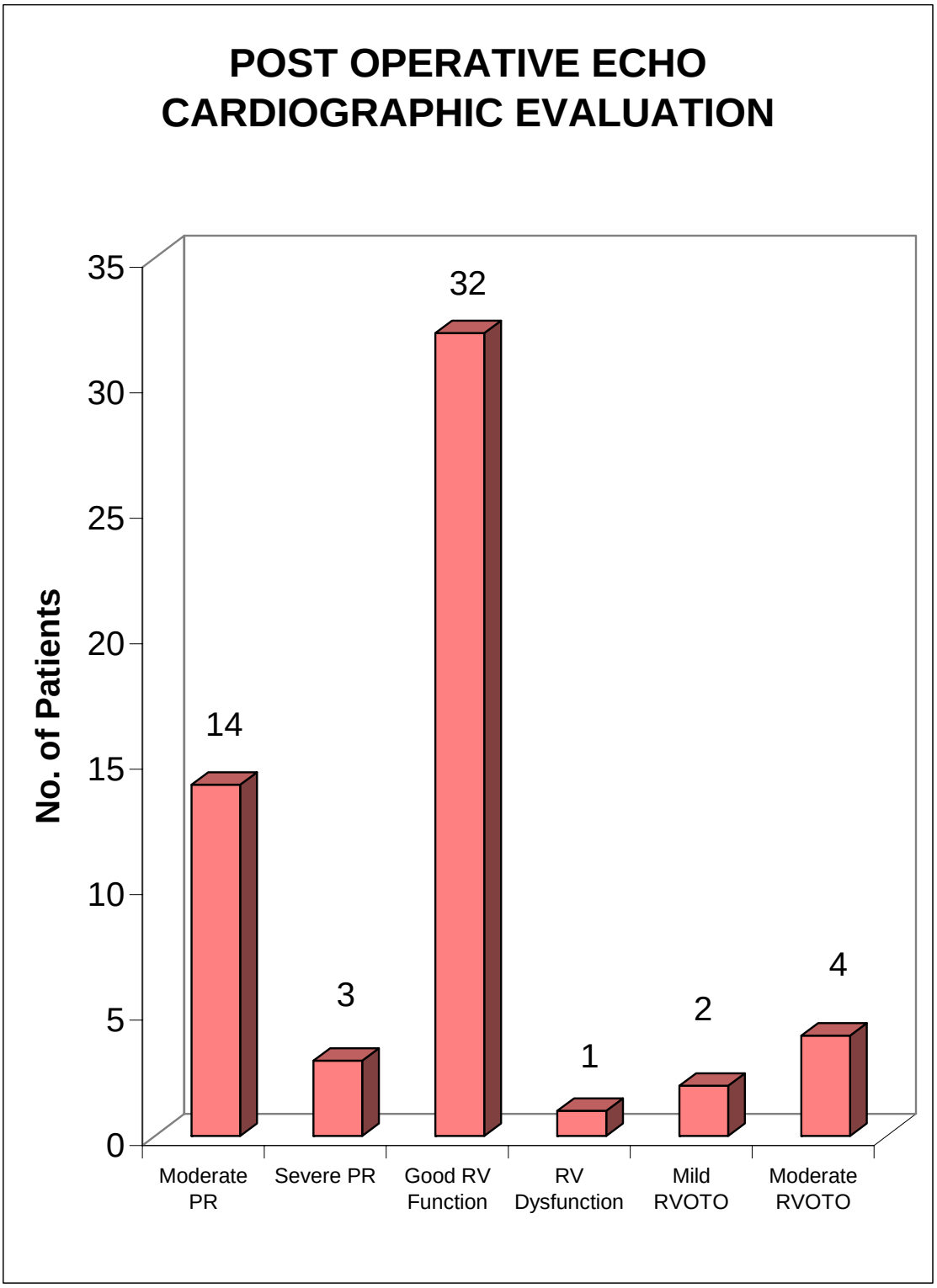

POST OPERATIVE ECHO
CARDIOGRAPHIC EVALUATION
No. of Patients
35
30
25
20
15
10
5
0
14
3
32
1
2
4
Moderate PR
Severe PR
Good RV Function
RV Dysfunction
Mild RVOTO
Moderate RVOTO

TABLE – 1

All patients were in sinus rhythm.2patients had residual insignificant shunts.6 patients had residual RVOT obstruction. Nollert and associates have reported use of a transannular patch to be associated with significantly worse operative and 1-year mortality as compared with those without a transannular patch. The free pulmonary regurgitation associated with a transannularpatch is thought to cause ventricular dilation and hamper ventricular function, especially during exercise. The results of our study show no significant difference in effort tolerance or significant difference in operative mortality and morbidity postoperatively between patients receiving a transannular patch /RVOTpatch and those without a patch. The studies on long term outcomes after total correction in adult TOF patients are limited. There was no difference in RV Ejection Fraction between patients receiving transannular patch and those without a patch. All of our patients were in sinus rhythm during the period of study. No arrhythmias were noted. All patients were taking Digoxin and Diuretics. Prehospital and Followup echo revealed no significant shunts, but residual RVOTO was found in 6 patients. RVOT GRADIENT was 20±5mmHg.Overall RV and LV functions were preserved.4 patients had mild to moderate LV dysfunction in the immediate postoperative period. One patient was

readmitted one year later with severe RV dysfunction and severe PR, severe ascites and poor general condition. He failed to improve with medical management and died.

The surgical correction of Tetralogy of Fallot has come a long way since 50 years. The objective of surgery at present is total correction at the age of 4 to 6 months. Despite the current shift in surgical strategy a significant number of patients present in the adulthood for surgery. Delayed presentation for surgery may be due to milder form of disease or delayed diagnosis or inaffordability of the patients for surgery. The management of adult TOF is a challenge because of the chronic hypoxia and patient is vulnerable to myocardial, neurologic, coagulation problems. Older age is also considered as an incremental risk factor for surgery, operative mortality and long term survival. In this context our study shows a considerable improvement in patient symptomatology with lower mortality and morbidity. Our Hospital mortality is 16%.Other studies show mortality as follows. Adil sadiq et al, SCTIMS show mortality rate of7%, Atik et al 5% ,Ditterech et al 16%.

HOSPITAL MORTALITY

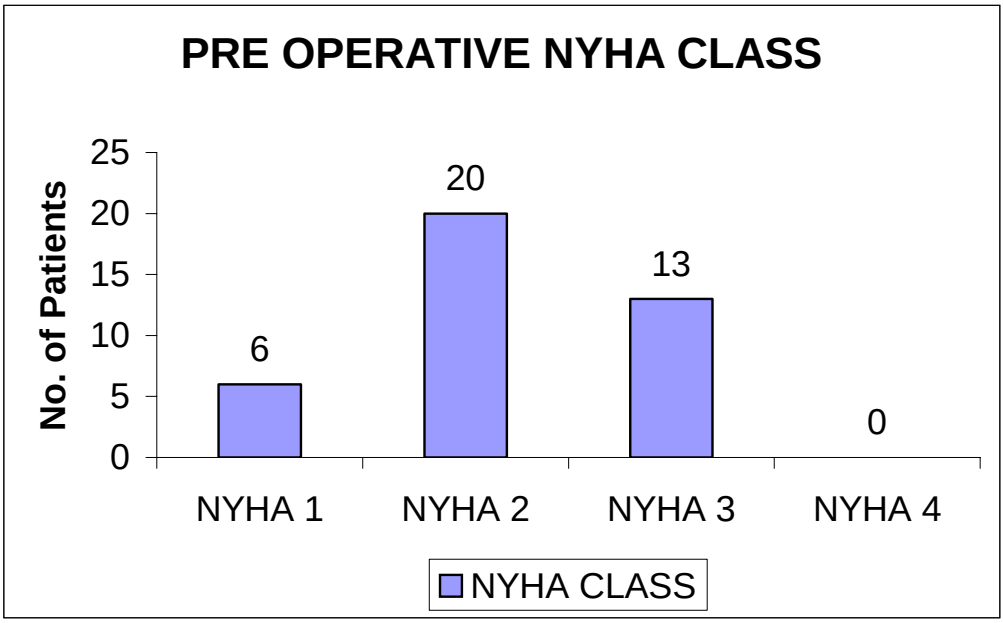
	Our series	Atik et al	AdilSadik et al	Ditterech et al
Percentage	16%	5%	7%	16%
No. of cases	6/39	3/39	4/58	3/9

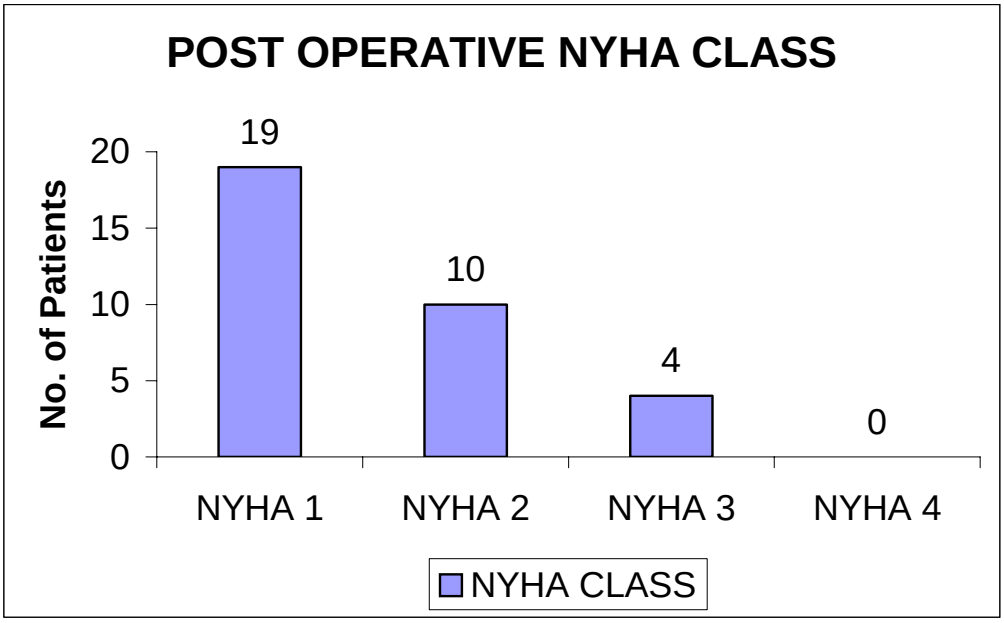
19 of our patients were in class 1, 10 in class 2 and 4 in class 3 postoperatively and on followup. AdilSadik et al showed 40 patients in class 1,5patients in class 2 and one patient in class 3 on followup of 46 patients. Atik et al showed one patient in class 1,10 patients in class 2 and one patient in class 3 on followup. TAB4

PRE OPERATIVE AND POST OPERATIVE

NYHA CLASS STATUS

OUR SERIES

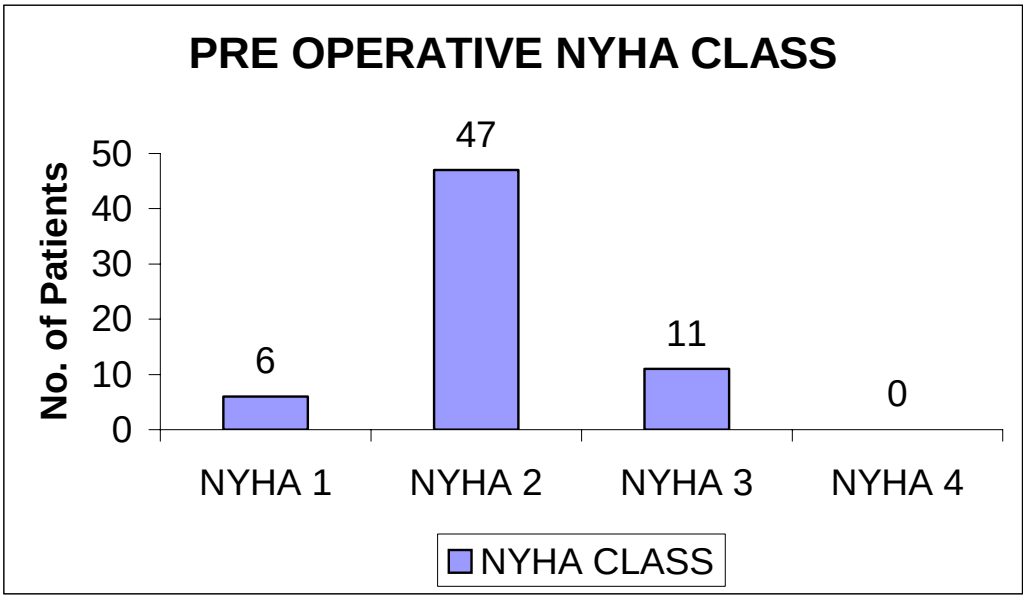


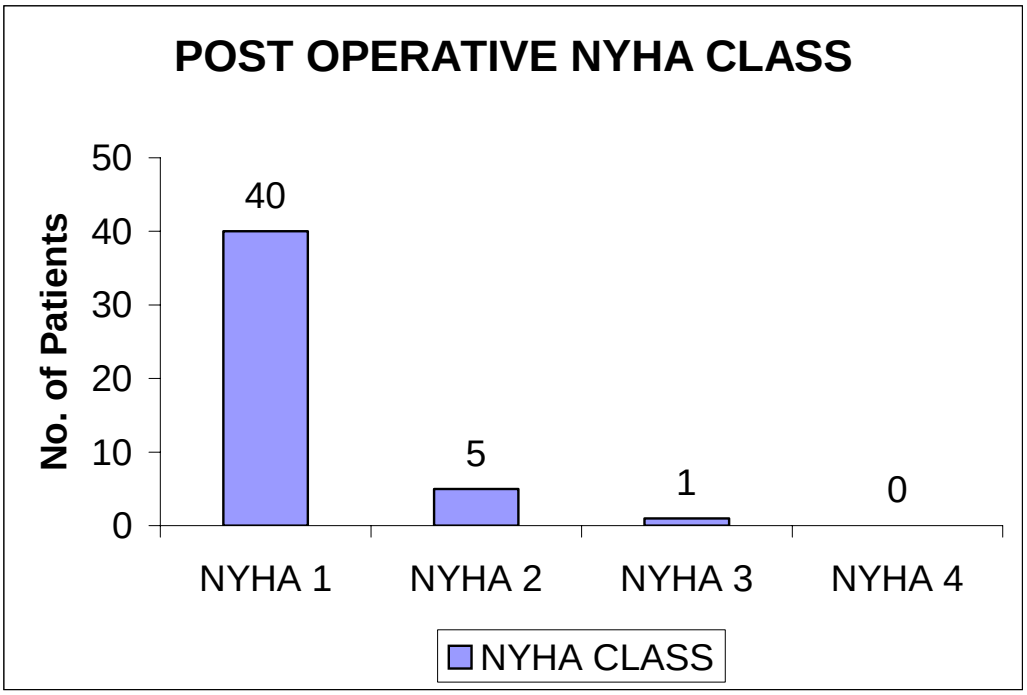


PRE OPERATIVE AND POST OPERATIVE

NYHA CLASS STATUS

ADIL SADIK ET AL

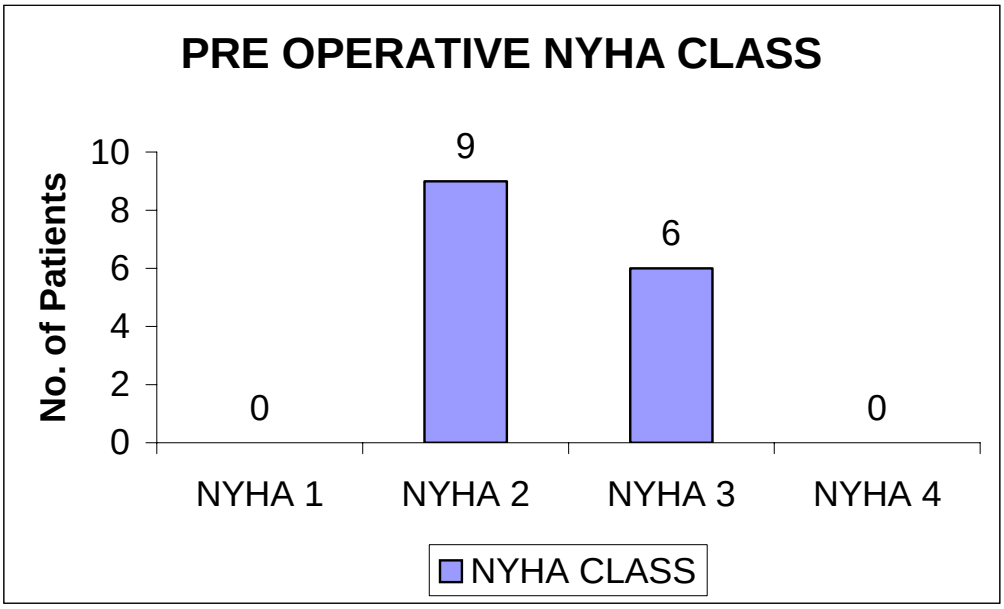


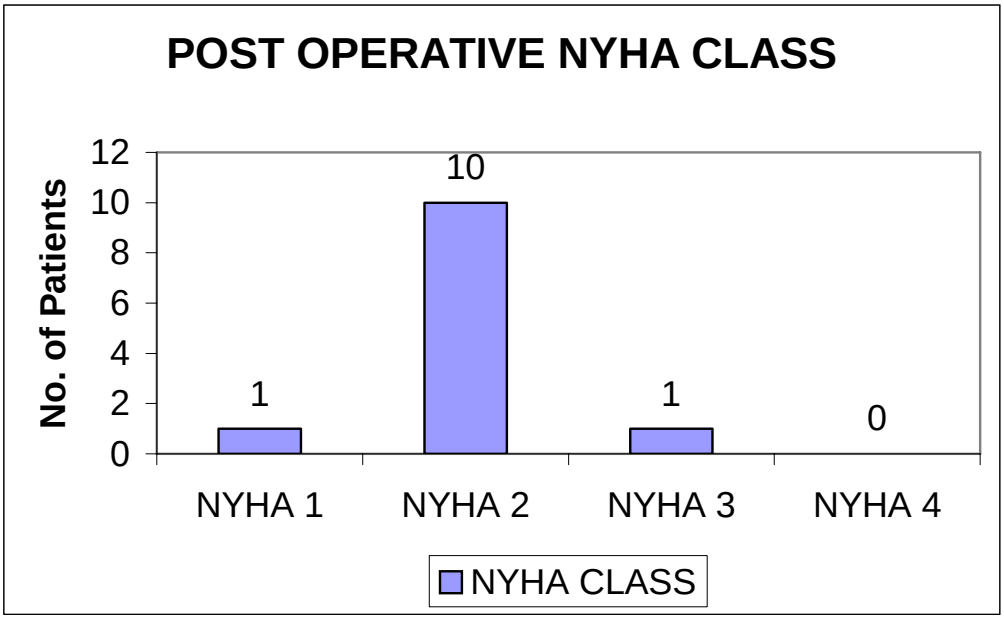


PRE OPERATIVE AND POST OPERATIVE

NYHA CLASS STATUS

ATIK ET AL





The preoperative mean RVOT gradient was 55-110 mmHg in our series. Atik et al 40-140mmHg,Adil Sadik et alSCTIMS 50-130 mmHg.

RVOT GRADIENT

Our series	Atik et al	AdilSadik et al
55-110 mmHg	40-140mmHg	52-130mmHg

.Intraoperative RV/LV pressure ratio after repair was 0.6-0.7.Adil Sadik et al showed 0.6±0.2.

RV/LV PRESSURE RATIO POST REPAIR

Our series	AdilSadik et al
0.6-0.7	0.6±0.2

The cause of death was low cardiac output and multiorgan dysfunction syndrome in 6 patients. 3 had postoperative bleeding. 40% patients had perioperative morbidity like prolonged ventilation and inotropic support, wound infection, elevated renal parameters .3 patients required readmission due to Right heart failure. One patient died during followup. There were no reoperations in the study period .Postoperative and followup echocardiographic evaluation showed no residual VSD. Residual stenosis seen in 6 patients.AdilSadik SCTIMS showed 3 cases of residual RVOTOin 39 patients.

RESIDUAL RVOTO

	Our series	**AdilSadik et al**
No. of cases	6/39	3/39
Percentage	15%	8%

Mild to moderate PR was seen in 14 patients. Adil sadiq et al SCTIMS ,study showed mild to moderate PR in 45 out of 54 patients.Atik et al showed mild to moderate PR in 9 of 39 patients. Transannular/RVOT patch was used in 22 of 39 patients in our study.

Atik et al used patch in 17of 39 patients . Adil sadiq et al used patch in 44 of 58 patients.

RVOT/TRANSANNULAR PATCHING

	Our series	**Atik et al**	**AdilSadik et al**	**Ditterech et al**
No. of cases	22	17	44	9
Total	39	39	58	19

All patients were in sinus rhythm .RV dysfunction was found in 13 of 39 patients (38%) in Atik et al series and our series showed preserved RV function. This may be attributed to the short duration of follow up in this study. Residual RVOTO was noted in 6 of 38 patients in our study. Bacha et al showed RVOTO in 8 of 57 patients. Adil sadiq et al showed 3 of 39 patients. 19 of our patients were in class 1, 10 in class 2 and 4 in class 3 on followup. None of the patients were cyanotic.

The management of adult tetralogy is a surgical challenge due to the chronic multisystem cellular hypoxia and compensatory polycythemia, which predispose theseatients to myocardial, neurologic, hematologic, and coagulation problems, as well as those related to the development of pulmonary collaterals .The hypertrophied and fibrotic right ventricular myocardium is prone to myocardial dysfunction and to ventricular arrhythmias. Transatrial repair is associated with a favorable outcome with lower mortality and morbidity. Patients have significant improvement in the quality of life and symptomatology. The need for reintervention remained very low and residual lesions requiring surgery like pulmonary valve replacement were not seen in the early followup. The major limitation in the study is the very brief period of observation. A longer followup as is done other comparable studies may throw light upon the long term effects of the surgical correction. It may be concluded that total single stage correction of TOF in adults may be undertaken with lower mortality, morbidity and significant improvement in functional status.

The first complete description of tetralogy of Fallot is credited to the French physician Etienne Fallot, who published his findings in 1888 (Fallot, 1888). It was not until 1945, however, that the first surgical treatment for tetralogy of Fallot was performed by Blalock at Johns Hopkins University (Blalock and Taussig, 1945).A number of innovative systemic-to-pulmonary shunt procedures were soon developed, followed by the first successful intracardiac repair using human crosscirculation. The first successful repair using a pump oxygenator was performed by Kirklin at the Mayo Clinic one year later. Numerous contributions have been made in the management of this defect since these initial pioneering efforts, including one-stage vs. two-stage repair, trans-annular patching, conduit repairs and transatrial repair, to name a few.

MORPHOLOGY

The classic components of the ‘‘tetrad’’ that comprise this defect are a ventricular septal defect (VSD), right ventricular outflow tract obstruction, aortic override and right ventricular hypertrophy. All of these individual components result from one basic morphological abnormality: anterior and left-ward displacement of the infundibular septum. The VSD in tetralogy of Fallot is a large, non-restrictive defect

that results from the malalignment of the left-ward or septal extent of the infundibular septum with the septal band (trabecula septomarginalis).Because the left-ward extent of the infundibular septum is displaced anterior to the anterior limb of the septal band, rather than between its anterior and posterior limbs, a large malalignment VSD results. The superior border of the VSD is the anteriorly rotated right-ward (parietal) extent of the infundibular septum. Posteriorly, it is bounded by the anteroseptal leaflet of the tricuspid valve. The inferior border is the posterior limb of the septal band, and the anterior border is the anterior limb of the septal band. The bundle of His penetrates at the posteroinferior edge of the defect. Although the VSD is generally subaortic in position, it may extend to the subpulmonary region when the infundibular septum is absent or deficient. Additional VSDs may exist in approximately 5% of patients and generally occur in the muscular septum.The anterior and left-ward displacement of the infundibular septum also results in right ventricular outflow tract obstruction.Hypoplasia of the right ventricular infundibulum, prominent muscle bands extending from the septal extent of the infundibular septum to the right ventricular free wall and contribute to the obstruction. A localized narrowing, or os infundibuli, frequently occurs at the inferior border of the infundibular septum.

The pulmonary valve is nearly always involved in the obstruction. The leaflets are usually thickened and tethered to the pulmonary arterywall. The pulmonary valve is bicuspid in 58% of patients, but it is the narrowest part of the outflow tract in a small minority of patients. The ‘‘annulus’’ of the pulmonary valve, although not a true fibrous structure, is nearly always smaller than normal. Important degrees of obstruction may also occur at the level of the right and left branch pulmonary arteries. Uncommonly, the left pulmonary artery may take origin from the ductus arteriosus and its intrapericardial portion may be completely absent. More commonly, localized narrowing of the origin of the right or left pulmonary arteries will be present and may be difficult to fully appreciate without special preoperative angiographic views. In extreme cases of anterior displacement of the infundibular septum, complete atresia of the distal right ventricular infundibulum and main pulmonary artery trunk may result. Pulmonary atresia is present in approximately 7% of patients with tetralogy of Fallot.Multiple aortopulmonary collateral arteries (MAPCAs) are usually found in those patients without an associated patent ductus arteriosus and provide a variable degree of the pulmonary blood flow. Unlike patients without pulmonary atresia, who possess centrally located areas of discrete pulmonary arterial stenoses, those with pulmonary atresia and MAPCAs

are more likely to have peripheral pulmonary stenoses. Approximately 5% of patients with tetralogy of Fallot will have complete absence of the pulmonary valve leaflets .Right ventricular outflow obstruction occurs at the level of the hypoplastic annulus and severe pulmonary regurgitation is generally present. Aneurysmal dilatation of the main pulmonary artery trunk and the right and left branch pulmonary arteries occurs and may result in compression of the distal tracheobronchial tree. The origin of the left anterior descending coronary artery from the right coronary artery occurs in approximately 3–5% of patients with tetralogy of Fallot .The left anterior descending coronary artery will cross the right ventricular outflow tract a short distance below the pulmonary valve annulus to reach the anterior interventricular septum and is susceptible to injury from an incision in this area.Rarely, a single right coronary artery gives rise to the left main artery which then crosses the right ventricular outflow tract. Major associated cardiac defects are relatively uncommon in tetralogy of Fallot. The most frequently associated lesions are atrial septal defect, patent ductus arteriosus, complete atrio-ventricular septal defect and multiple ventricular septal defects. Other less common defects include persistent left superior vena cava, anomalous origin of the left anterior descending coronary artery and aberrant origin of the right or left pulmonary artery.

DIAGNOSIS

The initial presentation of the patient with tetralogy of Fallot is dependent on the degree of right ventricular outflow tract obstruction. Most commonly, cyanosis is mild at birth and gradually progresses with age as the obstruction increases due to increasing hypertrophy of the right ventricular infundibulum. Cyanosis tends to become significant within the first 6–12 months of life in these patients. In such situations, the obstruction is entirely or predominantly at the infundibular level. In addition, these patients may develop characteristic cyanotic ‘‘spells’’, which are periods of profound systemic hypoxaemia. These spells are characterized by a marked decrease in pulmonary blood flow and an increase in the right-to-left shunt across the VSD, directing desaturated blood into the aorta.A smaller percentage of patients will present with significant cyanosis at or shortly after birth. In this group the outflow tract obstruction is nearly always due to a hypoplastic pulmonary valve annulus, with or without severe right ventricular infundibular obstruction or hypoplasia. Cyanosis is constant in these patients, due to the fixed nature of the obstruction to pulmonary blood flow. Patients with atresia of the pulmonary valve and main pulmonary trunk will be dependent on a patent ductus arteriosus or systemic aortopulmonary

collateral arteries for pulmonary blood flow. In the latter situation,the collateral arteries may be such that pulmonary overcirculation exists and congestive heart failure is present Cyanosis is the main physical finding. The characteristic systolic murmur results from the right ventricular outflow tract obstruction and is usually moderate in intensity. Typically the murmur disappears in the presence of a ‘‘spell’’. Continuous murmurs, best heard over the back and bilateral lung fields, will be heard in patients with a large amount of systemic aortopulmonary collateral artery flow. The characteristic electrocardiographic finding is that of right ventricular hypertrophy from pressure overload of the right ventricle. Right axis deviation will also befound. Left ventricular hypertrophy may be seen in those patients with increased pulmonary blood flow from large shunts or collaterals. On chest radiography, the heart size is generally normal and the pulmonary artery segment may be small. The aortic arch is right-sided in approximately 25% of patients. The characteristic ‘‘boot-shaped’’ heart results from elevation of the cardiac apex from the hypertrophied right ventricle and a concave upper left heart border caused by a narrow main pulmonary artery. The diagnosis is usually easily established by echocardiography. The typical malalignment VSD with aortic override and right ventricular outflow tract obstruction is well visualized. Often the location of the left anterior

descending coronary artery can be visualized as well. The peripheral pulmonary artery anatomy is poorly seen, however, and delineation of pulmonary artery anomalies constitutes one of the main indications for cardiac catheterization. The presence and precise anatomy of any significant aortopulmonary collateral arteries are determined by aortography. Branch right and/or left pulmonary artery stenoses, as well as non-confluent pulmonary arteries, must be sought by special cranial tilt views. Coronary arterial anatomy is seen following LV injection. Particular search is made for anomalous origin of the Left Anterior Descending artery(LAD) from RCA and for the rare but surgically important associated origin of LCA from the Pulmonary artery. Crossing the right ventricular outflow tract with the catheter may precipitate a spell and is generally not advised. In patients with long-standing cyanosis and those with pulmonary over circulation, measurement of the pulmonary artery pressure and pulmonary vascular resistance may also be required to exclude fixed pulmonary hypertension, which might preclude repair.

INDICATIONS FOR OPERATION

Most patients with tetralogy of Fallot have satisfactory systemic arterial oxygen saturation at birth and require no treatment. Progression of hypoxaemia will ultimately occur, and when the oxygen saturation falls below 75–80% operative intervention should be performed. Hypoxaemic spells may occur from transient reductions in pulmonary blood flow, due to a sudden increase in right ventricular outflow tract obstruction and a decrease in systemic vascular resistance. The occurrence of hypoxaemic spells is also generally considered an indication for operation, although in some cases medical management with propranolol may be used to delay operation. In those patients where specific indications have not yet developed, elective complete repair is recommended in most institutions by 1 year of age and, preferably by 3–6 months of age. Singlestage complete repair is being done in many centres, regardless of age but an initial systemic-to-pulmonary artery shunt procedure is preferred by others , particularly when symptoms occur within the first 6 months of life. The presence of pulmonary atresia, branch pulmonary artery hypoplasia, anomalous origin of the anterior descending coronary artery from the right coronary artery, or severe associated non-cardiac anomalies are other generally accepted

indications for a shunt rather than primary repair in the infant at several institutions. Benefits of early complete repair include normal growth and development of organs, elimination of hypoxaemia, less need for extensive right ventricular muscle excision, better late left ventricular function, decreased incidence of late dysrhythmias.Many other institutions, however, would still prefer primary repair, even when repair must be accomplished with the insertion of a conduit between the right ventricle and the pulmonary arteries. The need for a trans-annular patch because of significant hypoplasia of the pulmonary valve annulus was formerly considered a contraindication to complete repair in the infant, but this risk has now been neutralized. When an initial systemic-to-pulmonary shunt procedure is chosen as part of a staged repair, the classic or modified form of the Blalock Taussigprocedure is most commonly selected. The classic Blalock–Taussig procedure is performed on the side opposite the aortic arch (ipsilateral to the innominateartery) in order to allow the most favourable angle for the subclavian artery to reach the pulmonary artery without kinking. Although this anastomosis can be satisfactorily constructed in patients of any age or size many surgeons currently prefer the modified shunt in very small neonates because of the small size of the subclavian artery. In the modified procedure, an interposition polytetrafluoroethylene conduit

is placed between the subclavian and pulmonary arteries. A 4mm graft is generally preferred in neonates because early complete repair may be performed and larger shunts may result in congestive heart failure. The results achieved by this procedure have been excellent, with an extremely low shunt failure rate and an acceptable duration of palliation.In the presence of severe hypoplasia of the pulmonary arteries, palliation with a right ventricular outflow tract patch or conduit, leaving the VSD open, may be used . This promotes symmetrical pulmonary artery blood flow and reduces the likelihood of branch pulmonary artery distortion from a shunt procedure. However, in most patients with tetralogy of Fallot and pulmonary stenosis, the hypoplasia of the pulmonary arteries is due to the lack of pulmonary artery flow itself and responds promptly to procedures that increase this flow. Therefore, leaving the VSD open may result in severe congestive heart failure and pulmonary oedema from a sudden increase in pulmonary blood flow.Branch left pulmonary artery stenosis is well known to develop in some patients after repair and should be looked for during a routine follow-up. This is more likely to occur when repair is performed in the neonate, as the presence of a patent ductus arteriosus can mask the origin of the left pulmonary artery, which may become stenotic after the ductal tissue involutes. Palliation of the small or unstable neonate with

catheter-based approaches has been employed at some centres. Balloon dilatation of the pulmonary valve and stenting of ductus arteriosus can be utilized for stabilization prior to definitive operative intervention or in place of a systemic-to-pulmonary artery shunt. This approach, however, has not been widely accepted or utilized at this time.

SURGICAL TECHNIQUE

Anaesthesia

Monitoring lines include a femoral or radial arterial catheter and a central venous catheter (femoral or SVC). Additional peripheral intravenous lines and bladder catheter are placed. Nasal, cutaneous, and rectal temperature probes are utilized. Standard cardiac anaesthesia with high-dose narcotics and paralysis is employed. Care is taken to avoid significant changes in systemic and pulmonary vascular resistance that may precipitate a ‘‘spell’’ or increase the right-to-left (R–L)shunt.Should important hypoxaemia occur, α-agonist drugs are recommended to increase systemic vascular resistance and minimize the R–L shunt. A transoesophageal echocardiogram probe is placed after anaesthesia is induced. Additional intracardiac lines are placed at the termination of the procedure

Surgical Technique

A midline sternotomy incision is performed and the heart is exposed. The precise distribution of the coronary artery branches is confirmed and preparation is made for cardiopulmonary bypass. Little manipulation of the heart is done in order to avoid precipitating severe hypoxaemia from a ‘spell’’. Existing systemic-to-pulmonary artery shunts are exposed for subsequent interruption, although they may be dissected after cardiopulmonary bypass is established if the exposure is particularly difficult. Heparin is administered (300 U/kg). The aorta is cannulated just proximal to the level of the innominate artery and each vena cava is directly cannulated. Alternatively, a single cannula may be placed in the right atrium if the repair is to be performed using deep hypothermia and circulatory arrest. The latter approach may be utilized in patients < 2.0–2.5 kg in weight. In this situation, the VSD is closed under a brief period of circulatory arrest and the outflow tract repair is performed on cardiopulmonary bypass. Otherwise repair is done using continuous cardiopulmonary bypass, normal/low flows, the systemic temperature is lowered to 25–28◦C and the left ventricle is vented through the right superior pulmonary vein. All shunts are ligated and/or divided, the main pulmonary trunk and bifurcation (if branch stenoses

are present) are mobilized, and the ductus arteriosus is ligated. After the aortic cross-clamp is applied, the cardioplegic solution (30 ml/kg) is administered. Repeat doses (15 ml/kg) are administered at 20–30 minute intervals. The ASD and VSD are closed and the RVOTO is alleviated. The heart is de-aired via the left ventricular vent and the aortic air needle. The cross-clamp is removed and the trans-annular patch or RV–PA conduit, as necessary, are completed with the heart beating. A patent foramen ovale may be left open in neonates, particularly when post-repair haemodynamic residua remain, including pulmonary insufficiency and elevated right ventricular pressure. Because the neonate tolerates volume overload poorly, due to a non-compliant right ventricle and elevated pulmonary vascular resistance, oxygen delivery can be maintained by allowing a limited R–L atrial shunt at the expense of mild to moderate cyanosis. This is not necessary in older patients. In order to achieve a satisfactory repair, the surgeon must be completely familiar with all details of the anatomy, including the size and distribution of the branch pulmonary arteries, the nature and size of the pulmonary valve annulus (junction between the right ventricle and main pulmonary trunk), extent of the right ventricular outflow tract obstruction, coronary artery distribution, anatomy of the VSD, and the presence of any associated defects. After aortic cross-clamping, a right

atriotomy is made to assess the anatomy. Stay stitches are placed on the anterior and septal leaflet of the tricuspid valve. Gentle traction on these stitches will facilitate the exposure. If an atrial septal defect or patent foramen ovale is present, it is closed at this time. The anatomy of the VSD and right ventricular outflow tract obstruction is viewed through the tricuspid valve. A retractor placed anteriorly through the tricuspid valve and the stay stitches in the septal and anterior leaflet aid in the exposure of the distal outflow tract. When the repair is accomplished entirely through the right atrial approach, the outflow tract obstruction is approached first. A traction suture placed in the anterior and septal leaflets of the tricuspid valve aids exposure of the distal infundibulum. The position of the anterior margin of the VSD and the aortic valve leaflets are noted, and the parietal extent of the anterosuperiorly malpositioned infundibular septum is visualized. Invaginating the right ventricular free wall with a finger placed from outside the heart facilitates this exposure. Muscle trabeculations along the anterior limb of the septal band (trabecula septomarginalis) are divided on the right-angled dissector down to the level of the moderator band if necessary. When repair is performed in infancy, excision of the parietal extent of the infundibular septum (trabecula parietomarginalis) is rarely necessary and simple division of the obstructing muscle bundles is all that is

required. A pulmonary valvotomy can now be performed through the right atrial approach. If exposure is not adequate, a vertical incision is made in the main pulmonary artery, through which a pulmonary valvotomy may be performed. Valve leaflets may be mobilized and fused commissures divided all the way to the pulmonary artery wall. At this time, an assessment of the diameter of the pulmonary valve annulus is made by inserting calibrated dilators across the right ventricular outflow tract into the pulmonary artery trunk. The decision to place a trans-annular patch is made if the estimated post-repair RV/LV pressure is predicted to exceed 0.7. In this situation the main pulmonary artery incision is extended onto the right ventricular outflow tract across the pulmonary valve annulus. It can be kept quite short, extending only a few millimeters proximal to the annulus, as the infundibular obstruction has been adequately relieved trans-atrially. Whenever possible, this incision is placed directly through the anterior commissure of the valve to allow the pulmonary valve leaflets to remain functional and decrease the amount of pulmonary regurgitation. In some centres, the use of a monocusp outflow tract patch is preferred to prevent, or at least minimize, pulmonary regurgitation. Monocusp pulmonary valves, however, will only provide competence for a relatively short period of time, but may be particularly useful with

associated pulmonary artery hypoplasia, distal pulmonary obstruction or decreased ventricular function. Care needs to be taken when placing a right ventriculotomy that an aberrant origin of the LAD from the RCA is not injured. It is often possible to limit the incision in such a way that it remains superior to an anomalous LAD when a trans-atrial repair is done (hockey stick incision). A monocusp valve constructed of Gore-Tex, pericardium or homograft tissue may be placed in the outflow tract if pulmonary insufficiency is to be avoided. However, these valves provide only short-term competence and require a longer incision in the right ventricular infundibulum. In certain conditions, including hypoplastic branch pulmonary arteries, distal stenoses and poor ventricular function, the early pulmonary valve competence may confer an important haemodynamic advantage and improve the postoperative course. Closure of the VSD is accomplished from the transatrial approach, regardless of whether or not a transannular patch is needed, as that allows the ventricular extent of the incision to be minimized to the length necessary only for relief of obstruction, not for VSD exposure. Visualization of the VSD is generally adequate through the tricuspid valve and is even easier after dividing obstructing muscle bundles. A patch of polytetrafluoroethylene (Gore-Tex) is cut to the appropriate size and sutured to the right side of the septum, utilizing a continuous

suture technique. Suturing is commenced at the angle between the anterior and posterior limbs of the septal band (trabecula septomarginalis), directly opposite the perimembranous rim, and begun superiorly over the infundibular septum and aortic valve. The sutures are kept close to the aortic valve annulus itself in order to avoid residual defects in muscle trabeculations. This initial arm of the suture is brought into the right atrium by passing the needle through the anterior leaflet of the tricuspid valve at its junction with the ventriculoinfundibular fold. The other needle is then brought inferiorly, past the medial papillary muscle and under any chordae tendinae from the septal leaflet of the tricuspid valve, until the posteroinferior rim of the defect is reached. At this point, suturing must be done approximately 5mm away from the crest of the VSD itself and only on the right ventricular side. This is done in order not to injure the bundle of His, which penetrates the floor of the atrial septum in the apex of the triangle of Koch and runs adjacent to this margin of the VSD. Attaching the patch to the septal leaflet of the tricuspid valve away from the penetrating bundle completes suturing. The suture is tied over a pledglet on the right atrial side .Although the trans-atrial approach can be accomplished in the majority of patients undergoing repair of tetralogy of Fallot, even in infants, it is not suitable when the right ventricular outflow tract is diffusely hypoplastic. In these

situations, repair is best done with an outflow tract patch to enlarge the diameter of the infundibulum. Severe hypoplasia of the pulmonary valve annulus and main pulmonary trunk are common in this situation, and the outflow patch is then carried across the valve onto the pulmonary artery. It may be necessary to extend the patch, or tailor a separate patch, onto the left pulmonary artery in order to ensure that the obstruction is sufficiently relieved, because the main pulmonary trunk and origin of the left (or right) pulmonary artery branch are frequently hypoplastic. When an outflow patch is required, either for relief of severe infundibular and annular obstruction or for isolated enlargement of the pulmonary valve annulus alone (when the repair has been accomplished trans-atrially), it should be kept to the shortest length possible on the right ventricular aspect in order to avoid damaging right ventricular function. Upon completion of systemic re-warming, cardiopulmonary bypass is discontinued in the usual fashion and the haemodynamics are assessed for important residual lesions. The peak RV/LV pressure ratio is measured to ensure that significant residual outflow tract obstructiondoes not exist. If the post-repair RV/LV pressure is in excess of 0.7 and a trans-annular patch has not been placed, bypass is resumed and a patch is inserted across the pulmonary valve annulus. If a trans-annular patch has been placed, other causes of persistent elevation of

right ventricular pressure must be considered, including branch pulmonary artery stenoses, hypoplastic peripheral pulmonary arteries, residual VSD or residual infundibular obstruction. In the absence of these conditions, it may be assumed that the right ventricular hypertension will be well tolerated and will, in fact, improve over the next 24–48 hours .Often this elevation in right ventricular pressure results from dynamic right ventricular outflow tract obstruction, particularly when an outflow patch is avoided, as in the case of a trans-atrial repair. Administration of an ultra-short acting β-blocking agent, such as Esmolol, can help to differentiate dynamic vs. fixed residual right ventricular outflow tract obstruction intraoperatively.

SPECIAL CIRCUMSTANCES

Pulmonary Artery Abnormalities

Stenosis of the origin of the left and/or right pulmonary arteries is frequently encountered in patients with tetralogy of Fallot. Left pulmonary artery stenosis is best augmented with placement of a separate patch. Simple extension of the right ventricular outflow patch onto the left pulmonary artery can cause flow disturbances, as well as distortion of the left pulmonary artery takeoff, due to the acute posterior

course which the vessel follows. Stenosis at the origin of the right pulmonary artery is more difficult to repair, because of the right angle that this vessel takes from the main pulmonary artery and the more difficult exposure resulting from the overlying ascending aorta. In this situation, a separate patch may be necessary to enlarge the proximal right pulmonary artery. Alternatively, resection of the stenotic area, if it is relatively localized, with end-to-end anastomosis using absorbable suture, may also give good results. Bifurcation stenoses involving both pulmonary artery origins may be repaired with a resection and end-to-end anastomoses of both pulmonary arteries. Alternatively, each branch of a bifurcated pulmonary artery allograft may be anastomosed to the distal pulmonary arteries beyond their stenoses. Less commonly, one of the branch pulmonary arteries may have an anomalous systemic arterial origin. Usually it is the left pulmonary artery that arises from a normally positioned ductus arteriosus and proceeds directly to the hilum of the lung without entering the pericardium. Exposure of the left pulmonary artery is best accomplished by first dissecting under the arch of the aorta to isolate the ductus. The pulmonary artery can then be followed toward the hilum to gain sufficient length for primary anastomosis to the side of the main pulmonary artery trunk. When the right pulmonary artery is not in continuity with the left, it is likely to originate from a right-sided

ductus off the innominate artery. Rarely, it may take origin from the descending aorta. When the branch pulmonary arteries are non-confluent, or confluent but with a significant stenosis centrally (usually at the insertion of the ductus itself), palliation with a systemic-to-pulmonary artery shunt is a poor choice of treatment. In these cases the shunt flow will be predominantly to one lung, while the other receives little or no flow. Reconstruction of the central pulmonary artery bifurcation using one or more of the techniques outlined above is preferred in order to provide symmetric pulmonary blood flow. This is best combined with complete intracardiac repair, although placing a shunt to the reconstructed pulmonary arteries may also be done The postoperative care of the patient with tetralogy of Fallot can be challenging. A right atrial pressure line is routinely placed for monitoring of volume status and as a guide to right ventricular function. Occasionally, particularly when left ventricular function is compromised, the addition of a left atrial monitoring line is also useful. Significant volume resuscitation is often necessary to provide adequate filling of the hypertrophied and non-compliant right ventricle. Inotropes are instituted to maintain adequate cardiac output and perfusion pressure once filling pressures are optimized. A patent foramen ovale, if present, is generally left open in the neonate to allow for R–L shunting, to

maintain cardiac output at the expense of mild systemic arterial desaturation in the early postoperative period. Atrial and ventricular pacing wires are placed for the diagnosis and management of postoperative arrhythmias. Junctional ectopic tachycardia (JET) is a relatively common postoperative arrhythmia and is treated with surface cooling, overdrive pacing and intravenous amiodarone administration.In addition to arrhythmias, important postoperative complications include tamponade, residual outflow tract obstruction, residual VSD, tricuspid insufficiency, and phrenic nerve palsy. Even small residual VSDs are poorly tolerated by a left ventricle not accustomed to volume overload, and tamponade may cause haemodynamic compromise early in the presence of a dilated right ventricle, as seen with pulmonary insufficiency. Tricuspid insufficiency may occur secondary to the sutures used for VSD patch placement and is poorly tolerated when pulmonary insufficiency and elevated right ventricular pressure coexist.

RESULTS

The early (hospital) mortality after repair of tetralogy of Fallot is currently 1–5% in most reported series (Groh, 1991; Castaneda, 1990; Gustafson *et al*., 1988;Karl *et al*., 1992). A review of 399 patients undergoing complete repair for tetralogy of Fallot (excluding repairs

performed concomitant with or subsequent to unifocalization of MAPCAs) at the University of Michigan between January 1993 and November 2004 show an overall early hospital mortality rate for complete repair of tetralogy of Fallot as 3% (95% confidence interval 1.2–4.6%).Repair within the first year of life did not influence the early outcome, as had been reported in a number of earlier series. The neutralization of this risk factor is largely due to improved intraoperative techniques (particularly the avoidance of excessive right ventricular outflow tract muscle excision), cardiopulmonary bypass management and refinements in postoperative care.Among 814 patients undergoing complete repair at the University of Alabama at Birmingham, survivorship at 1 month and at 1, 5 and 20 years was 93%, 92%, 92%and 87%, respectively (Kirklin *et al*., 1989). Survival was slightly less than that for an age-, race- and gendermatched control population. In their analysis, the risk factors for late death were older age at repair, high peak right/left ventricular pressure immediately after repair (> 0.85) and the presence of a Potts shunt. The use of a trans-annular patch was not found to be a risk factor for premature late death.

LATE COMPLICATIONS

The most common indications for reoperation are the result of long-term complications of the right ventricular outflow tract, such as severe pulmonary regurgitation,residual outflow tract obstruction or conduit failure. Less commonly, reoperation is necessary for a residual VSD. Even relatively small residual defects are poorly tolerated after tetralogy repair, and reoperation is recommended when the pulmonary:systemic flow ratio exceeds 1.5. The most common location for a residual VSD is at the posteroinferior margin of the patch, presumably because suturing in this area is done superficially to avoid heart block.

Additional reasons for reoperation include pulmonary valve stenosis, left pulmonary artery stenosis and aortic valve insufficiency. The exact amount of residual outflow tract obstruction required for reoperation is controversial, but when right ventricular pressure exceeds 60 mm Hg, relief of residual obstruction is generally indicated. An earlier review from the University of Michigan of tetralogy of Fallot patients undergoing complete repair in the first month of life demonstrated a 1 month, 1 year and 5 year freedom from reoperation rates of 100%,93% and 63% (Hennein *et al*., 1995). The relatively high

reoperation rate in this age group, however, was influenced by the complex anatomy associated with symptomatic neonates with tetralogy of Fallot, including a high incidence of non-confluent pulmonary arteries,branch pulmonary artery stenoses and conduit insertion.An intraoperative pressure ratio between the right and left ventricles, at the time of initial repair, of 0.75 or greater is an independent risk factor for reoperation.The precise surgical intervention depends on the nature and location of the obstruction, but a careful search for branch pulmonary stenosis should be done. When the obstruction is located at the annular level, a trans-annular patch should be inserted. Overall survival and functional status following reoperation is very good, with a 10 year actuarial survival of 92%, with 93% of these patients in a New York Heart Association classification of I or II.

When significant residual pulmonary artery branch stenoses are diagnosed late after repair, treatment by balloon dilatation and/or stent insertion can be very effective. Although the effect of pulmonary insufficiency on early and late mortality has been neutralized in recent series, long-standing pulmonary insufficiency appears to have certain deleterious effects on ventricular function and exercise capacity. This is particularly true when additional residual lesions are present, such as a

VSD or peripheral pulmonary stenosis. In a series of 20 patients with isolated pulmonary insufficiency studied an average of 9 years after repair, showed significantly lower right and left ventricular ejection fractions when compared with a group of patients with competent pulmonary valves.

This was found despite the fact that all the patients analysed in this study were asymptomatic and had excellent haemodynamic repairs. Thus, it is likely that some patients with pulmonary insufficiency will develop symptoms many years after repair, even in the absence of other significant residual defects. When symptoms develop, the insertion of a pulmonary valve has been shown by a number of groups to improve functional status and ventricular function.

Following reoperation, a statistically significant drop in RV/LV pressure ratio has been demonstrated,along with a reduction by at least one NYHA classification grade. The indications for pulmonary valve replacement in the absence of symptoms of exercise intolerance or congestive heart failure are not well defined, but this procedure should not be considered in the presence of poor ventricular function, tricuspid insufficiency and progressive right ventricular dilatation. Early reoperation for asymptomatic right ventricular dysfunction improves the

chance for full recovery of ventricular function, as well as decreasing the prevalence of ventricular arrhythmias (Ilbawi *et al*., 1987). A review of patients undergoing elective pulmonary valve replacement has demonstrated that the operative risk was low (1.1%). In addition, functional status was NYHA I for 90% of patients following repair, with a 10 year survival of 95%

Interest has again developed in the use of monocusp valves in the right ventricular outflow tract. The objective is to minimize pulmonary regurgitation and thereby reduce the long-term effects on the right ventricle. Short term data has failed to consistently demonstrate improved postoperative recovery in patients with monocusps vs.those with standard trans-annular patches, particularly when the trans-annular patch is kept short and right ventricular muscle excision is avoided . Additionally, longer-term follow-up has shown a failure of monocusps to maintain satisfactory function after a few months.

- PATIENT NAME:
- AGE/SEX:
- MRD NO:

- ADDRESS:
- OCCUPATION:
- CONSULTANT IN-CHARGE:
- UNIT
- DATE OF ADMISSION:
- DAE OF SURGERY:
- DATE OF DISCHARGE:
- CHIEF COMPLAINTS:
- CARDIAC SYMPTOMS: SHORTNESS OF BREATH/EFFORT INTOLERANCE/PALPITATIONS/PND/CHESTPAIN/PEDAL EDEMA/RESPIRATORY DISTRESS:
- NEUROLOGIC SYMPTOMS:
- CONSTITUTIONAL SYMPTOMS: FEVER/ARTHRALGIA
- PAST HISTORY/FAMILY HISTORY
- GENERAL EXAMINATION:
- PALLOR +/-
- CYANOSIS +/-
- CLUBBING +/-
- PEDAL EDEMA
- PR/BP:
- SYSTEMIC EXAMINATION
- CARDIOVASCULAR SYSTEM
- NEUROLOGICAL EXAMINATION
- INVESTIGATIONS
- Hb/PCV/ESR:
- ECG:
- CHEST X-RAY:
- TRANS-THORACIC ECHOCARDIOGRAPHY:

- CARDIAC CATHETERISATION:
- CORONARY ANGIOGRAPHY:
- CARDIAC CT/MRI
- DIAGNOSIS:
- SURGICAL PROCEDURE:
- APPROACH: TRANSATRIAL/INFUNDIBULAR RESECTION/VSD CLOSSURE:
- TRANSANNUALAR PATCHING/RVOT PATCHING
- CROSS CLAMP TIME:
- TOTAL CPB TIME:
- OUTCOME:
- POST OP VENTILATORY SUPPORT/INOTROPIC SUPPORT:
- POST OP ECHO/FOLLOWUP ECHO RESIDUAL VSD/RVOT GRADIENT/PR/RV FUNCTION.

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